Vision: Cerebral Pathways and Disease

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Today's Talks

- Start with a patient (video)
- Overview of the visual system
  - Retina to occipital cortex (V1)
  - Beyond visual cortex
- Unusual visual problems
- David Leopold: Blindsight

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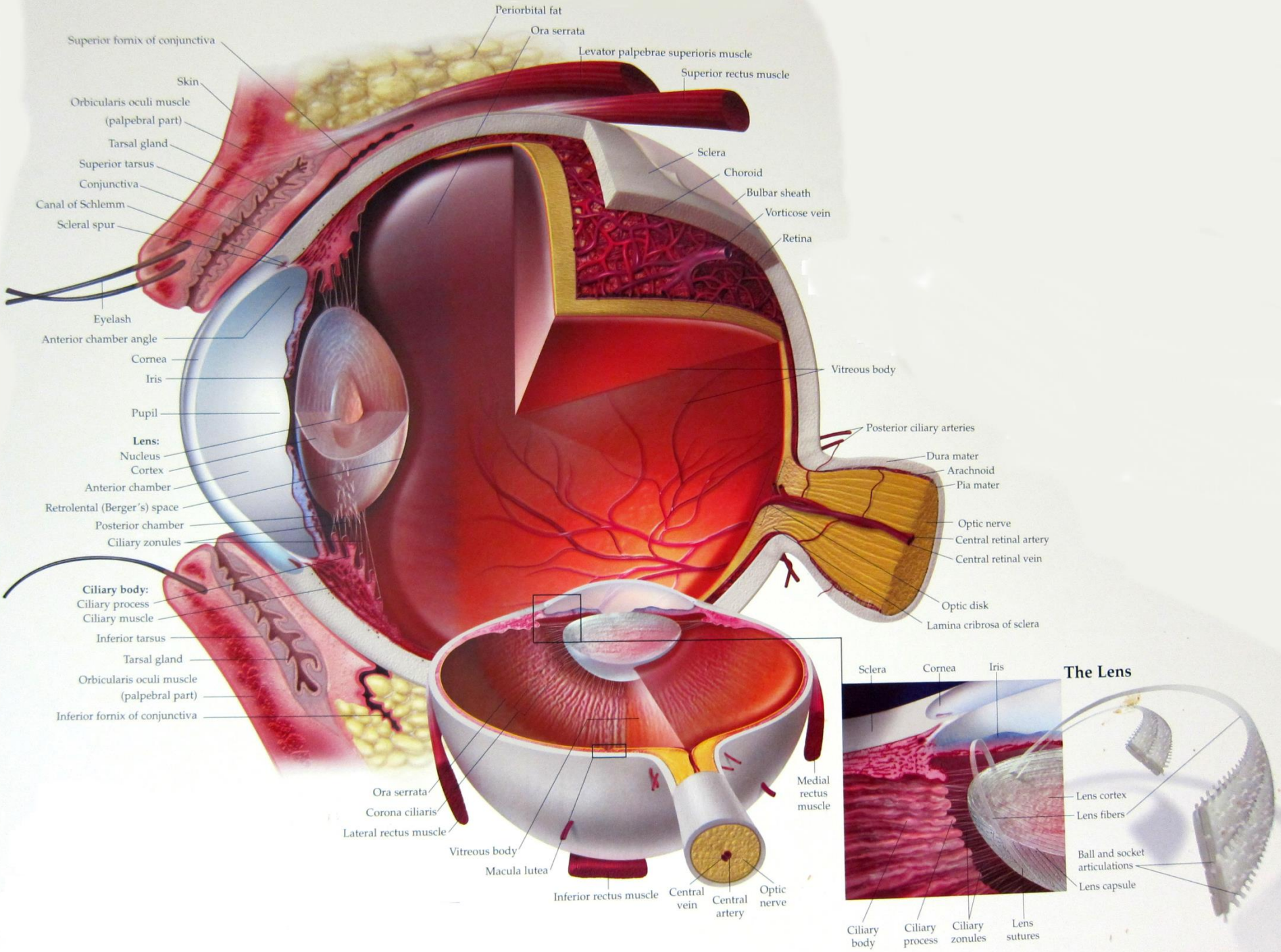
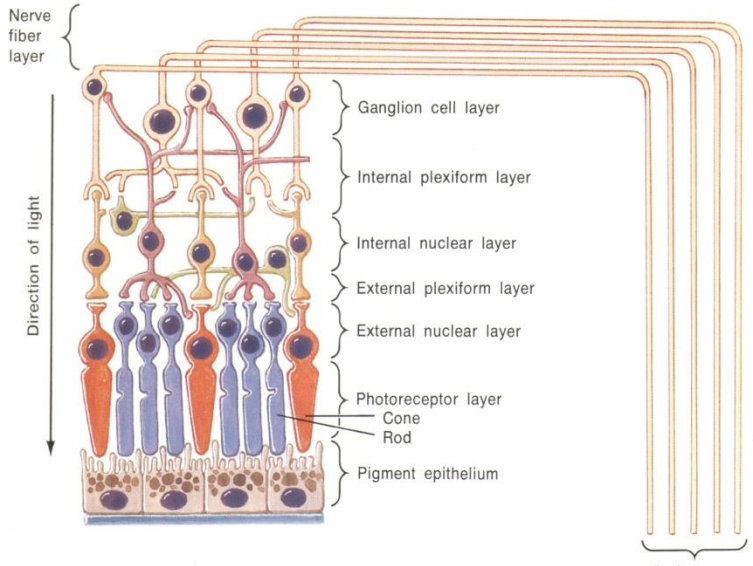
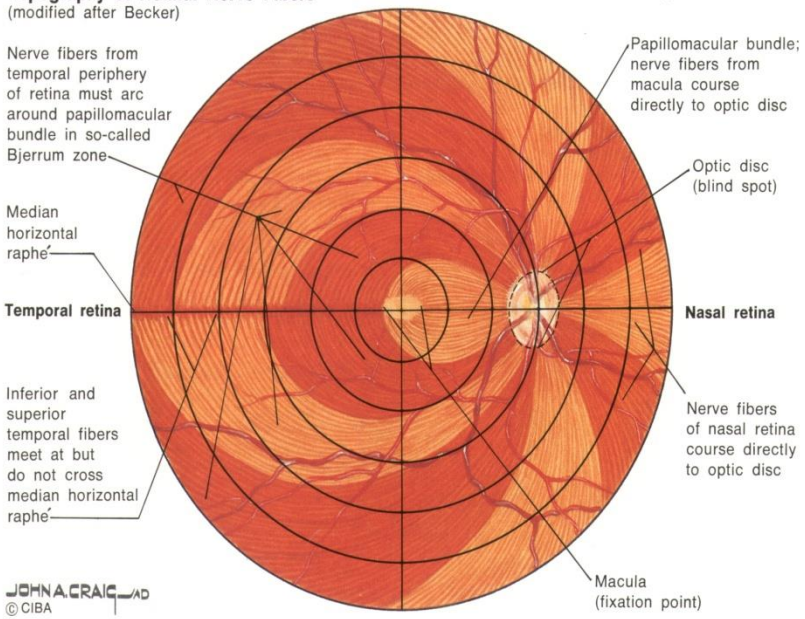

Periorbital fat
Ora serrata
Superior fornix of conjunctiva
Levator palpebrae superioris muscle
Superior rectus muscle
Skin
Orbicularis oculi muscle
(palpebral part)
Tarsal gland
Superior tarsus
Conjunctiva
Canal of Schlemm
Scleral spur
Sclera
Choroid
Bulbar sheath
Vorticose vein
Retina
Eyelash
Anterior chamber angle
Cornea
Iris
Pupil
Lens:
Nucleus
Cortex
Anterior chamber
Retrolental (Berger's) space
Posterior chamber
Ciliary zonules
Vitreous body
Posterior ciliary arteries
Dura mater
Arachnoid
Pia mater
Optic nerve
Central retinal artery
Central retinal vein
Optic disk
Lamina cribrosa of sclera
Ciliary body:
Ciliary process
Ciliary muscle
Inferior tarsus
Tarsal gland
Orbicularis oculi muscle
(palpebral part)
Inferior fornix of conjunctiva
Ora serrata
Corona ciliaris
Lateral rectus muscle
Vitreous body
Macula lutea
Inferior rectus muscle
Central vein
Central artery
Optic nerve
Medial rectus muscle
Sclera
Cornea
Iris
Ciliary body
Ciliary process
Ciliary zonules
Lens sutures
The Lens
Lens cortex
Lens fibers
Ball and socket articulations
Lens capsule

Schema of Retinal Neurons
(modified after Polyak)



Topography of Retinal Nerve Fibers
(modified after Becker)



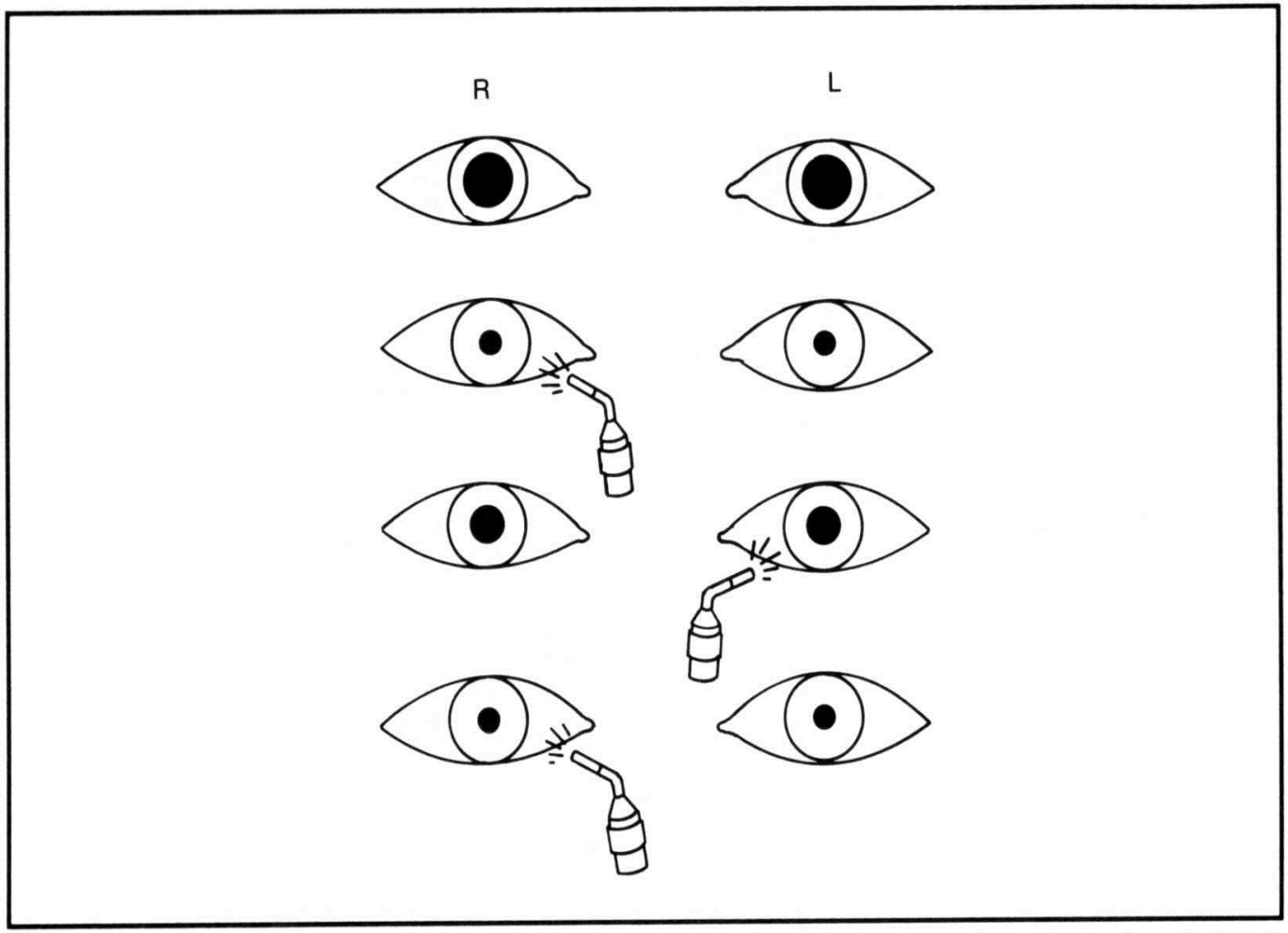
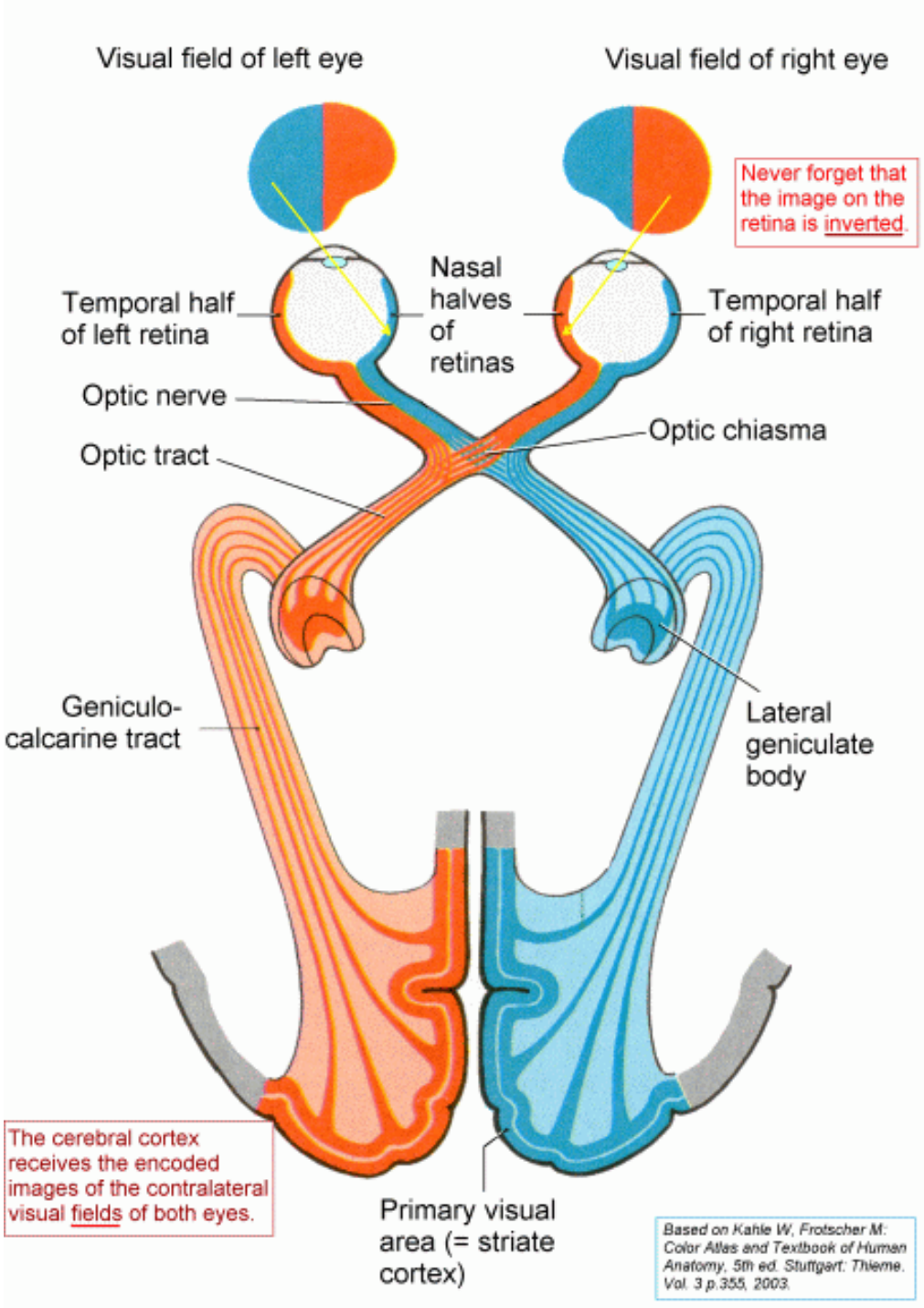


Figure 8-5. Afferent pupillary defect in left eye using swinging flashlight test. The pupils constrict when the light is shined in the right eye; however, when the flashlight is swung back to the left eye, both pupils dilate.


Visual field of left eye
Visual field of right eye
Never forget that the image on the retina is inverted.
Temporal half of left retina
Nasal halves of retinas
Temporal half of right retina
Optic nerve
Optic chiasma
Optic tract
Geniculo-calcarine tract
Lateral geniculate body
The cerebral cortex receives the encoded images of the contralateral visual fields of both eyes.
Primary visual area (= striate cortex)
Based on Kahle W, Frotscher M: Color Atlas and Textbook of Human Anatomy, 5th ed. Stuttgart: Thieme. Vol. 3 p.355, 2003.

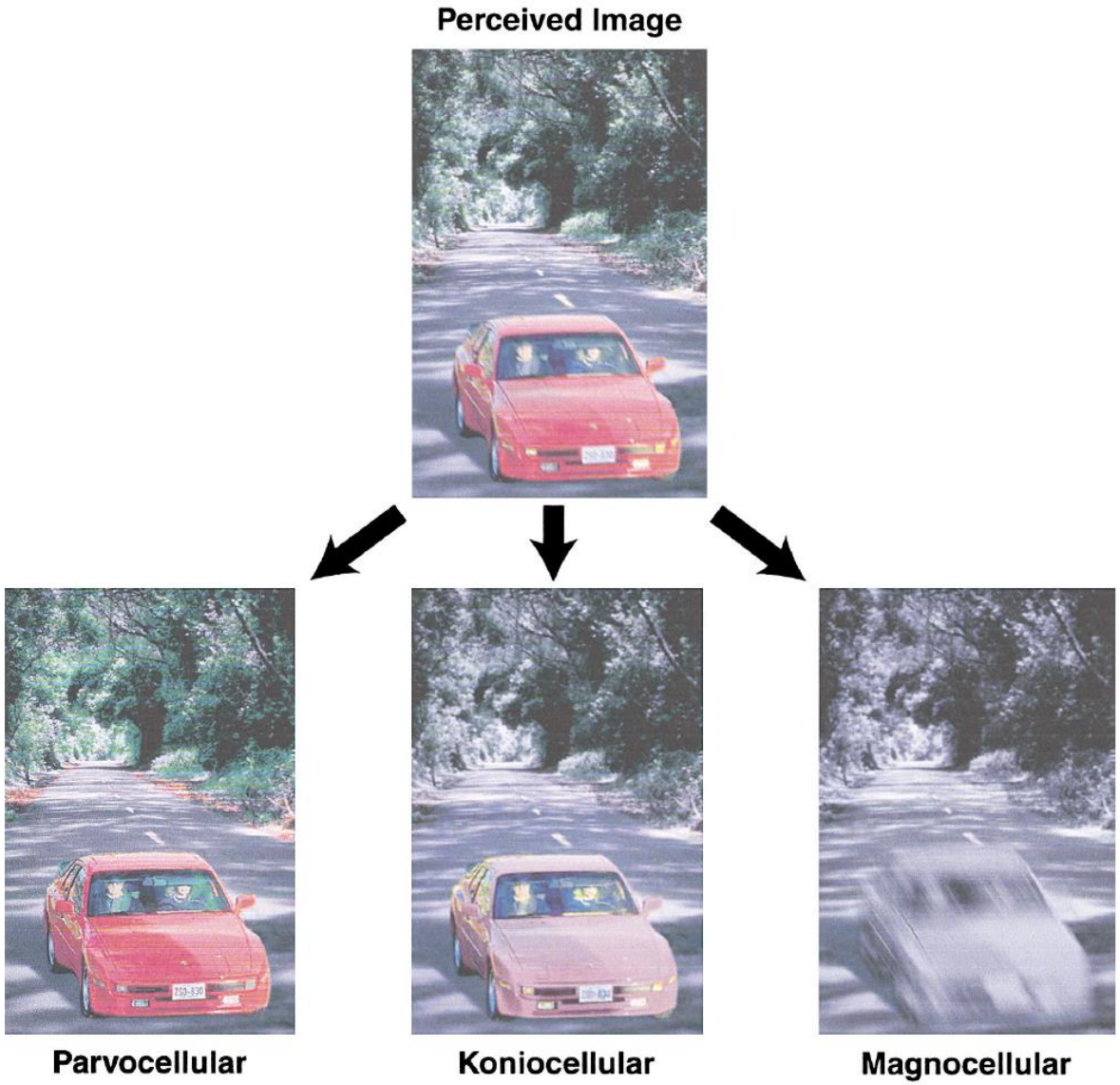


Fig. 1. Visual information that is used to create the perceived image of a visual scene (*top* photograph) is conveyed to the lateral geniculate through three pathways. The lower photographs were manipulated to simulate the characteristics of the perceived image that are conveyed along each of these pathways. The parvocellular pathway (*lower left* photograph) conveys fine spatial detail and is thought to be wavelength selective primarily for red and green opponency. The koniocellular pathway conveys information concerning blue-yellow opponency and low spatial detail (*lower middle* photograph). The magnocellular pathway (*lower right* photograph) conveys color-blind information of low spatial detail, sensitive to motion. (Artist: David Fisher.)

From: Girkin & Miller SurvOphthalmol 45(5) Mar-Apr 2001

Lateral Geniculate

- 6 layers
- Crossed contralateral fibers in lamina 1, 4 and 6, uncrossed ipsilateral fibers in 2, 3 & 5.
- Large magnocellular (M) fibers carrying motion, form and monochromatic brightness information in lamina 1 and 2.
- Small parvocellular (P) fibers carrying color (red-green), shape, texture and depth information in lamina 3, 4, 5, 6.
- Intercalated koniocellular carrying blue cone information.

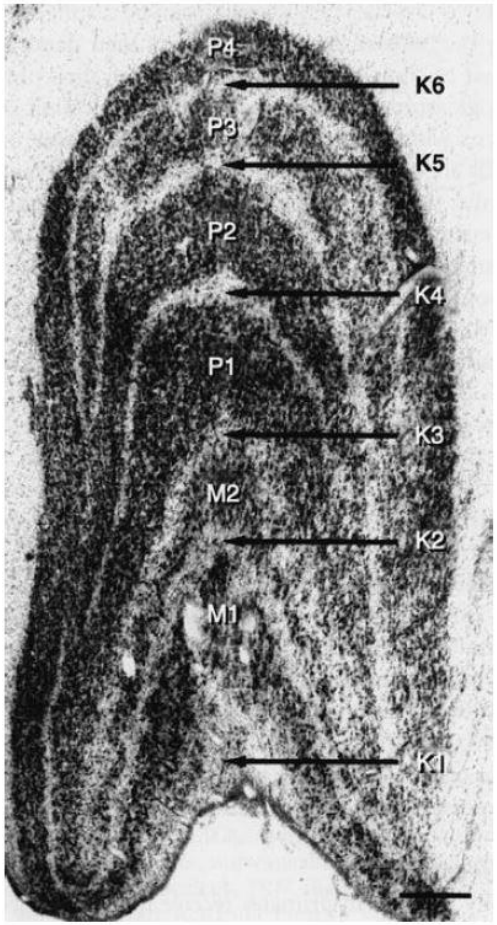
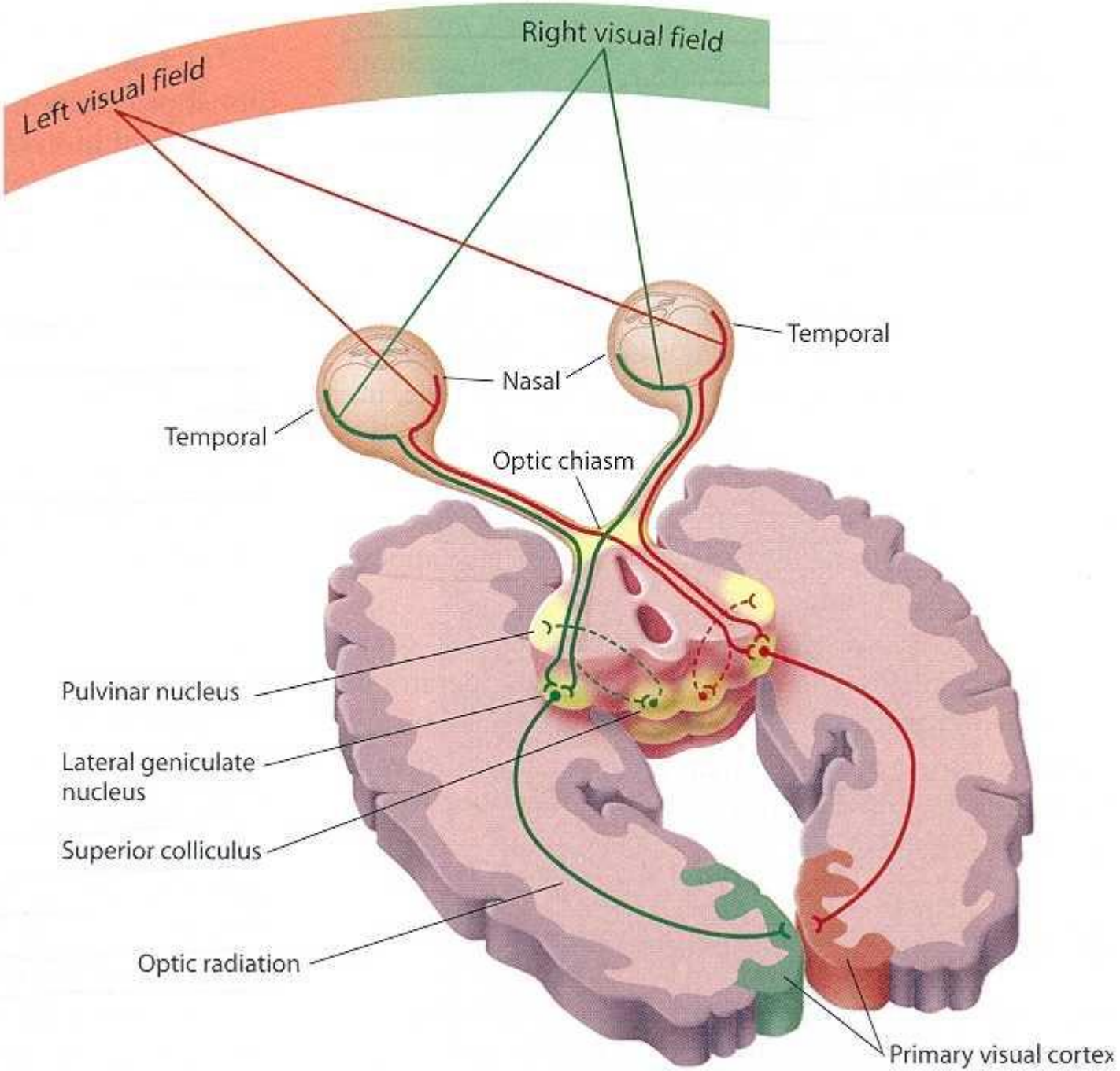
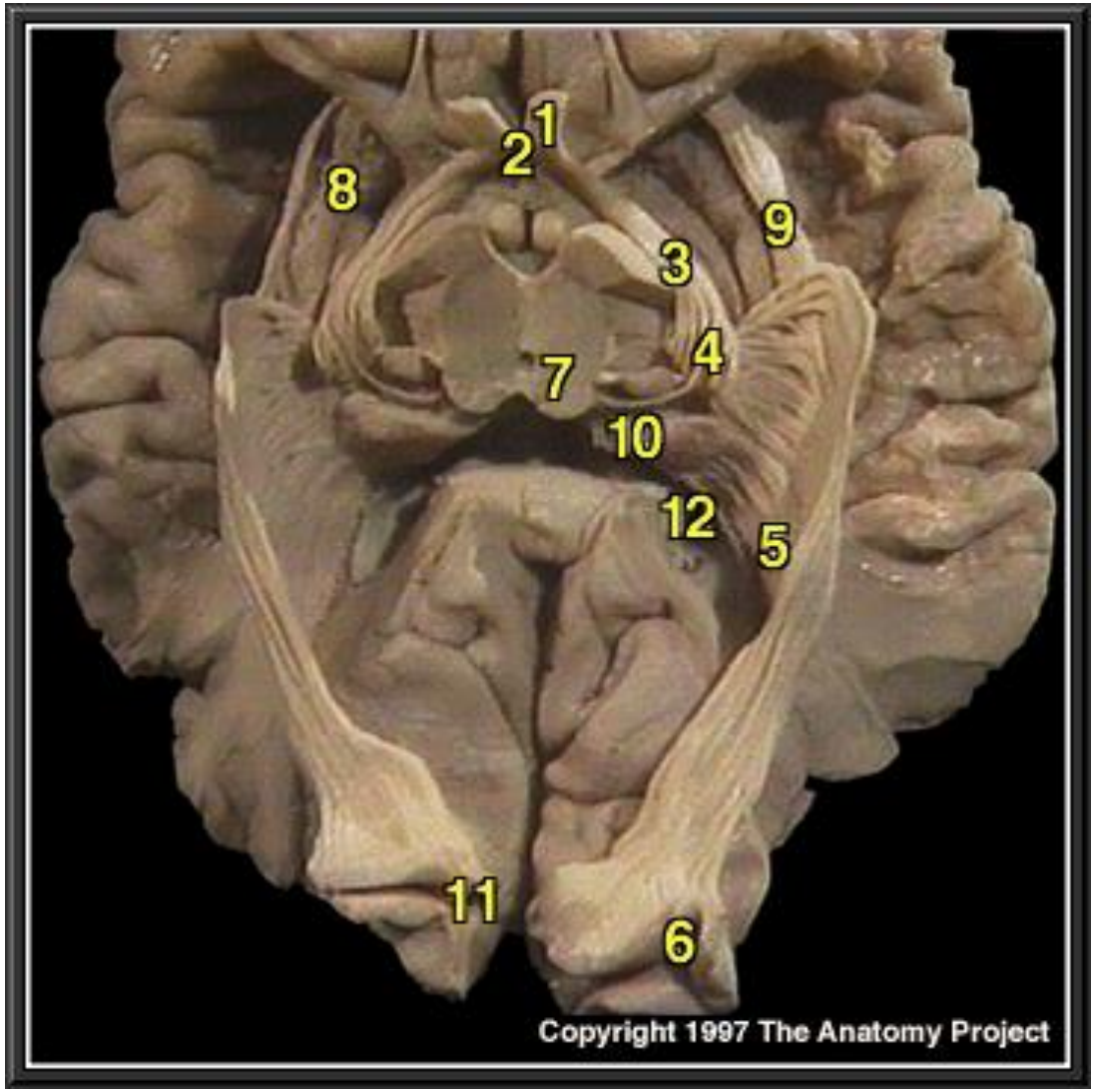


Figure 1.48. A coronal section through the lateral geniculate nucleus of a macaque monkey showing the parvocellular (P), magnocellular (M), and koniocellular (K) layers. At this plane there are four P layers, two M layers, and six K layers. Laminae 1, 4, and 6 receive input from the contralateral eye; laminae 2, 3, and 5 receive input from the ipsilateral eye. The thin layers ventral to each of the six primary lamina to which the K-retinal ganglion cells project are known as ''intercalated'' or interlaminar layers. Scale bar = 500 μm. (From VA Casagrande, JM Ichida. The lateral geniculate nucleus. In: Kaufman PL, Alm A, eds. Adler's Physiology of the Eye. 10th ed. St Louis, Mosby, 2003:657.)


Left visual field
Right visual field
Temporal
Nasal
Temporal
Optic chiasm
Pulvinar nucleus
Lateral geniculate nucleus
Superior colliculus
Optic radiation
Primary visual cortex



1- Optic nerve
2- Optic chiasma
3- Optic tract
4- Lateral geniculate body
5- Optic radiation
6- Visual cortex
7- Superior colliculus
8- Putamen
9- Inferior occipitofrontal fasciculus
10- Pulvinar of the thalamus
11- Calcarine fissure
12- Posteroinferior horn of the lateral ventricle

Localization: Where's the Problem?

- Visual Fields
  - Confrontation
  - Static threshold
  - Kinetic
- Possibilities:
  - Prechiasmal
  - Chiasmal
  - Retro-chiasmal

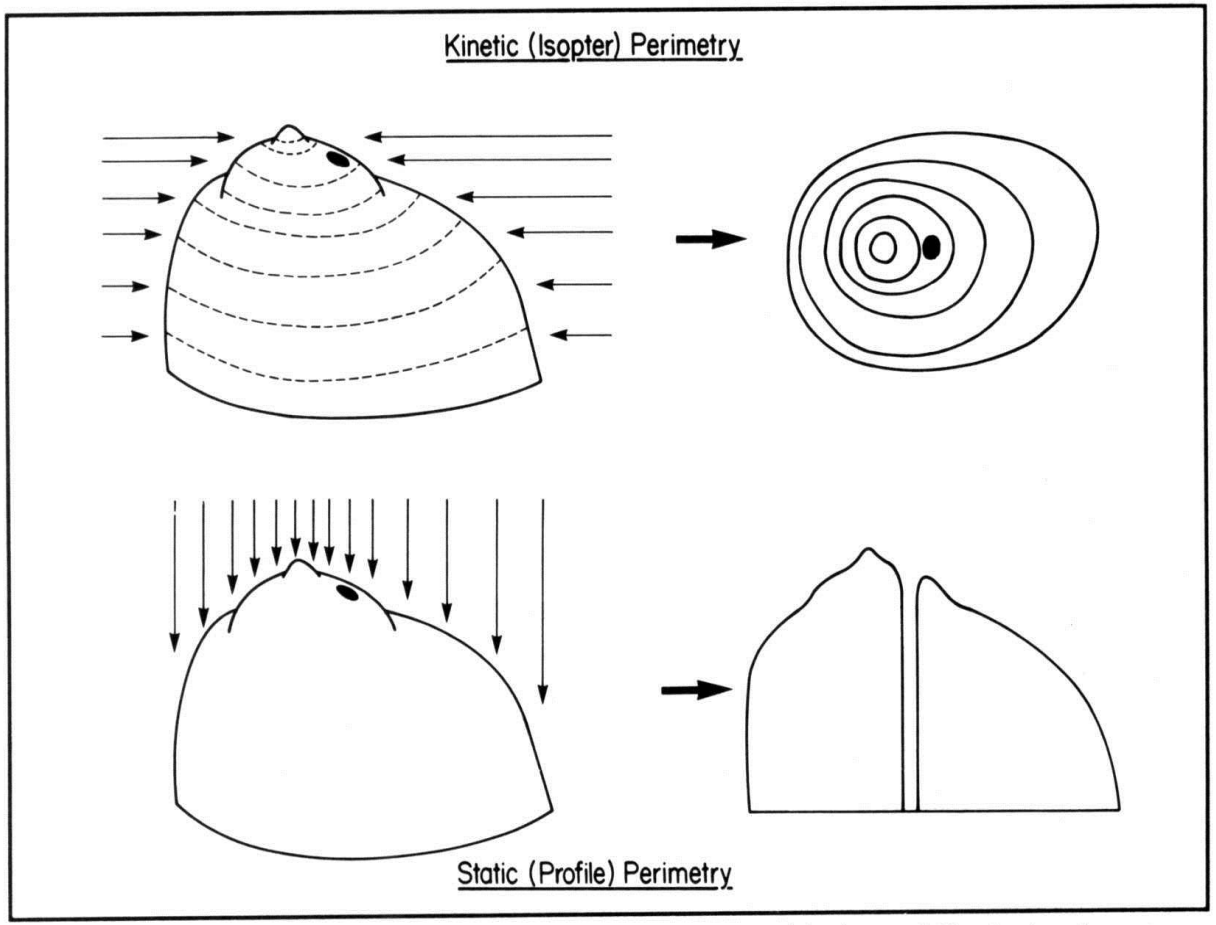


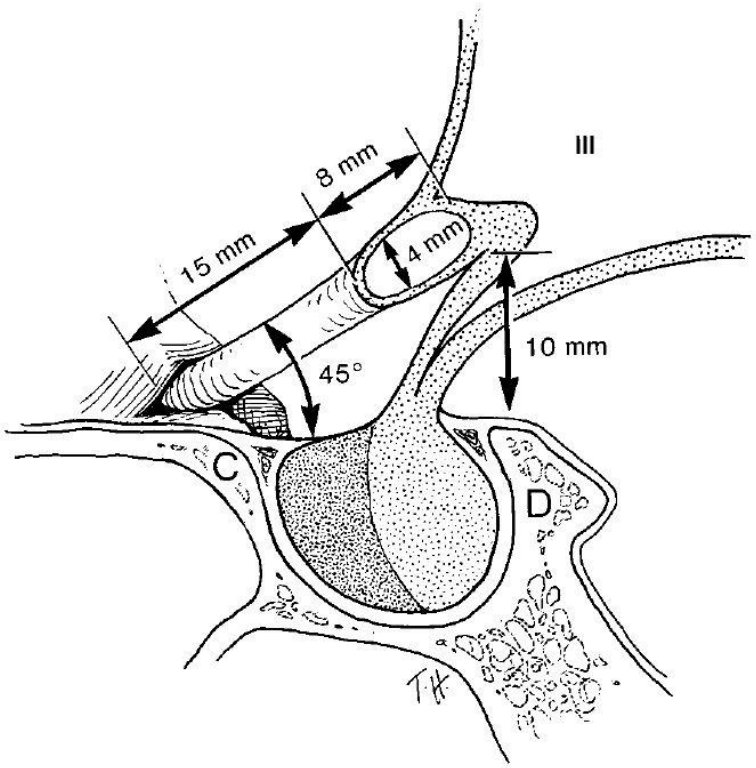
Figure 1-2. Kinetic perimetry: each set of target size-color-intensity and background illumination determines a different level of the island being tested and results in a different oval-shaped cross-section or isopter; note the six isopters at right, the result of testing six levels of the island. Static Perimetry: the target is held stationary at different points along the selected meridian; the intensity of the target is slowly increased until it is detected by the patient; the intensity required determines the upper level (the greatest sensitivity) of the island at this point.

Pre-chiasmal

- Vision loss in one or both eyes but visual fields tend to respect horizontal meridian and are incongruous (not the same shape in each eye).
- The problem could be possibly due to:
  - cornea, lens (refractive)
  - retina (diabetic retinopathy, retinitis, RP…)
  - optic nerve head (glaucoma, papilledema, ION,…)
  - optic nerve (MS, compression, inflammation,…)

Chiasmal

- Vision loss in both eyes – bitemporal (outer visual field loss). Respects vertical meridian.



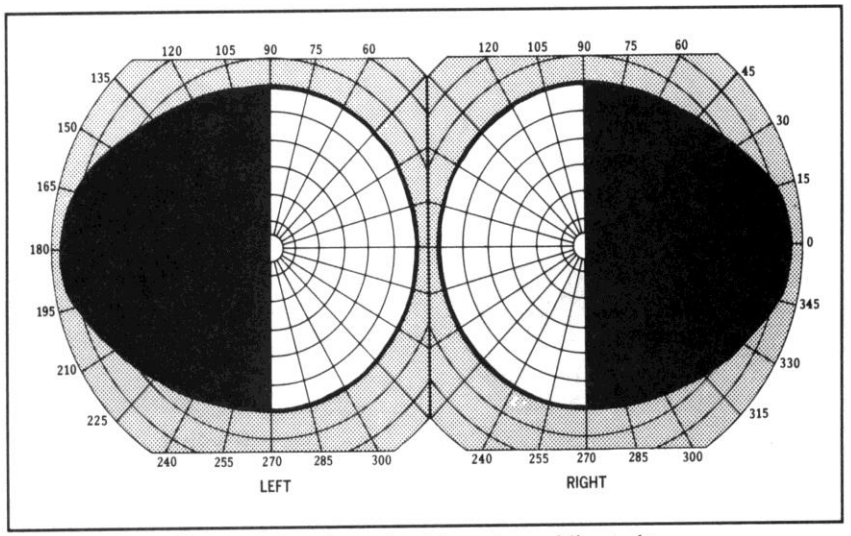


Figure 1-9. Bitemporal hemianopia due to interruption of decussating nasal fibers in chiasm.

Figure 1.43. Relationships of the optic nerves and optic chiasm to the sellar structures and 3rd ventricle (III); C, anterior clinoid; and D, dorsum sellae. (Redrawn from Glaser JS. Neuro-ophthalmology. Hagerstown, MD, Harper & Row, 1978.)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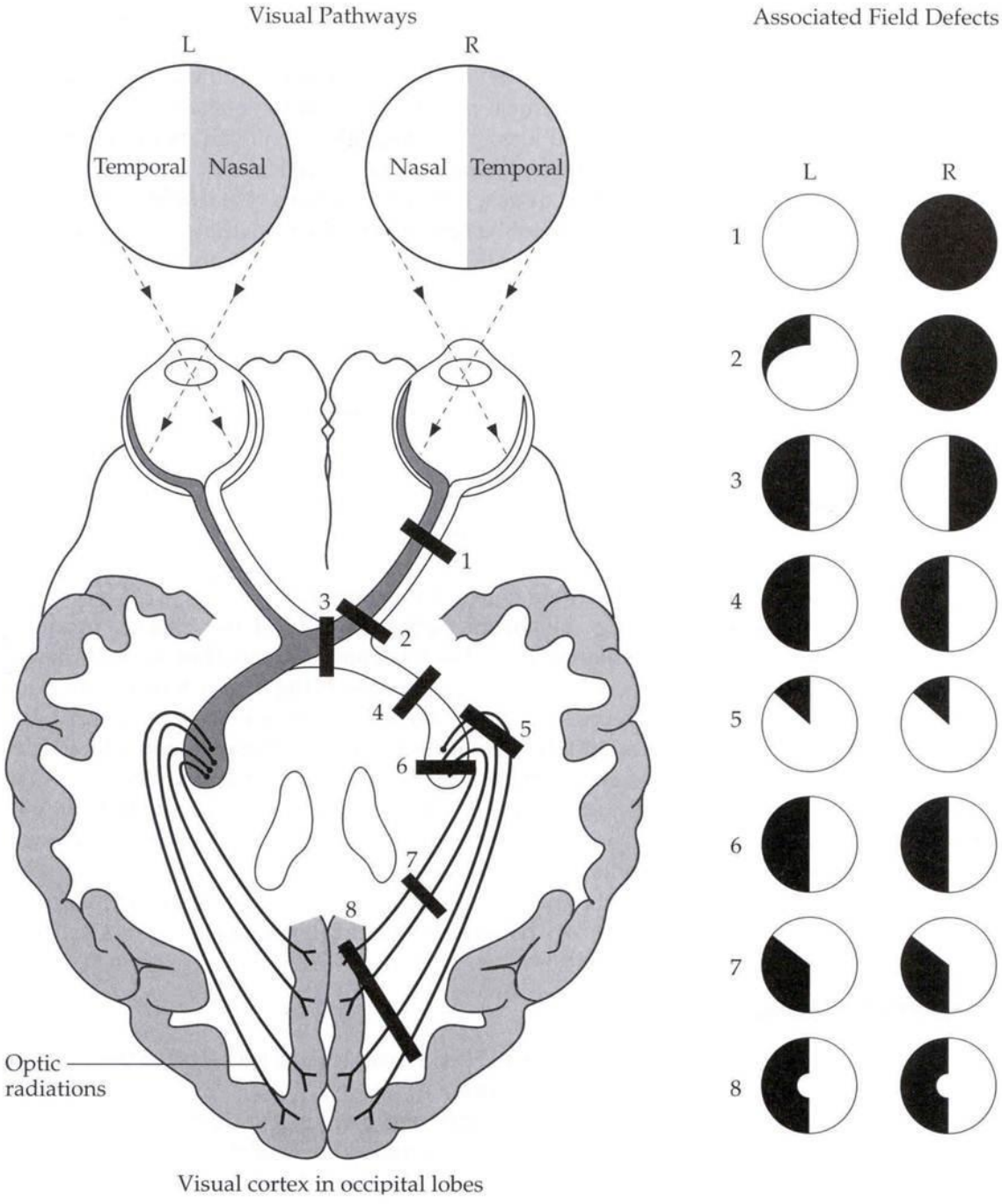
Retro-chiasmal

- Vision loss on same side (right or left half) in both eyes. Respects vertical meridian.
  - Right visual field loss –> left brain involvement
  - Superior vision loss -> inferior brain involvement
  - More congruous –> more posterior involvement


Visual Pathways
L
R
Temporal
Nasal
Nasal
Temporal
1
2
3
4
5
6
7
8
Optic radiations
Visual cortex in occipital lobes
Associated Field Defects
L
R
1
2
3
4
5
6
7
8

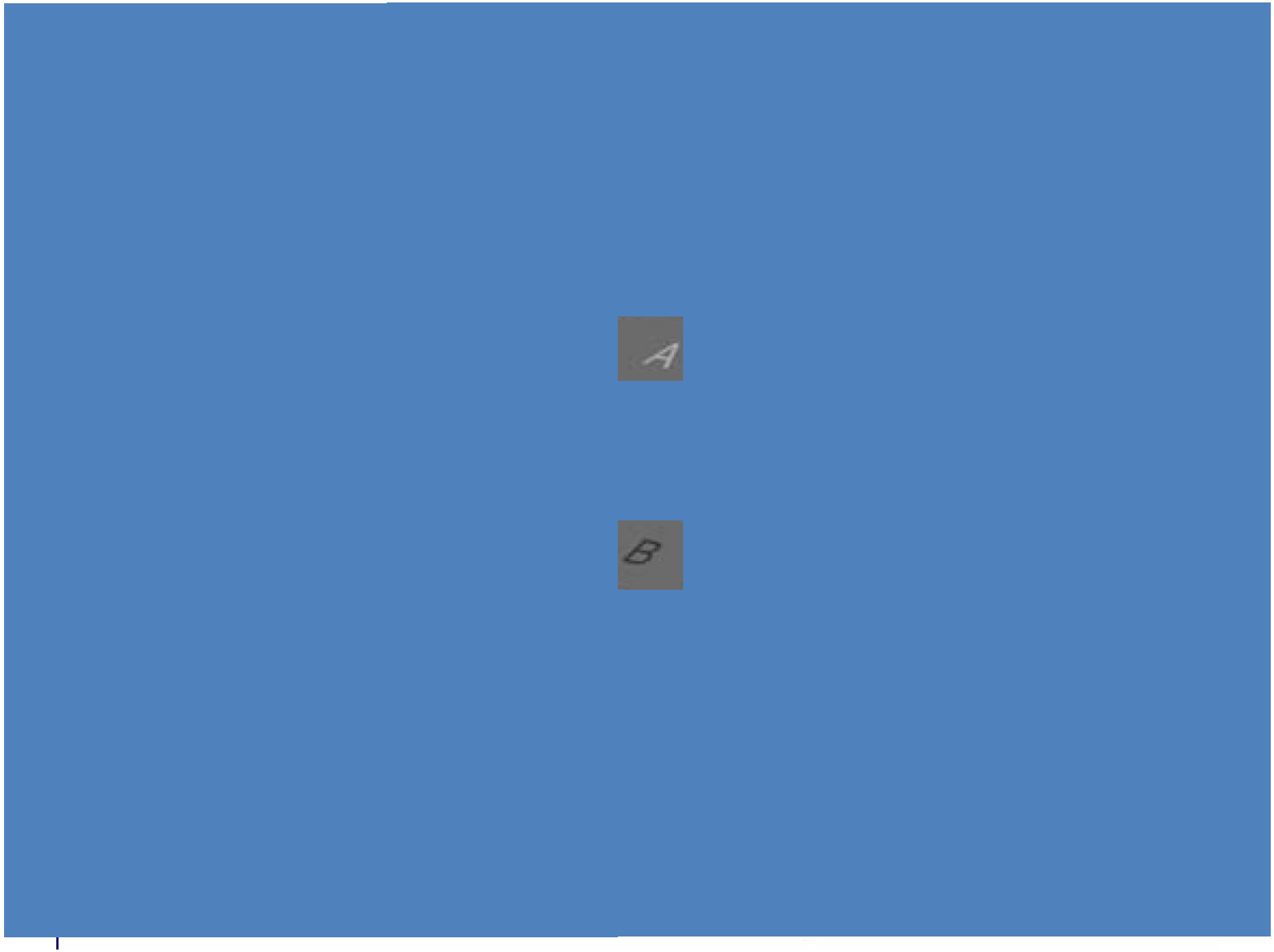
Diseases Leading to Visual Pathways Vision Loss

- Stroke
- Tumor – brain or metastasis, benign, malignant
- Inflammation – Vasculitis, Multiple sclerosis, …
- Degeneration - leukodystrophy, Alzheimer's, …
- Other (AVM, trauma, …)

- **Diagnosis:**
  - Neuroimaging (MRI, CT, Angiography)
  - Company it keeps

Vision is Complex

- Contrast and color constancy
- We foveate yet have a perception of space as being constant
- Illusions can sometimes help us notice what's going on behind the scenes
- Two examples (& that dress controversy)


A
B

Change Blindness (using flicker)

(from J. Kevin O'Regan -- http://nivea.psycho.univ-paris5.fr)

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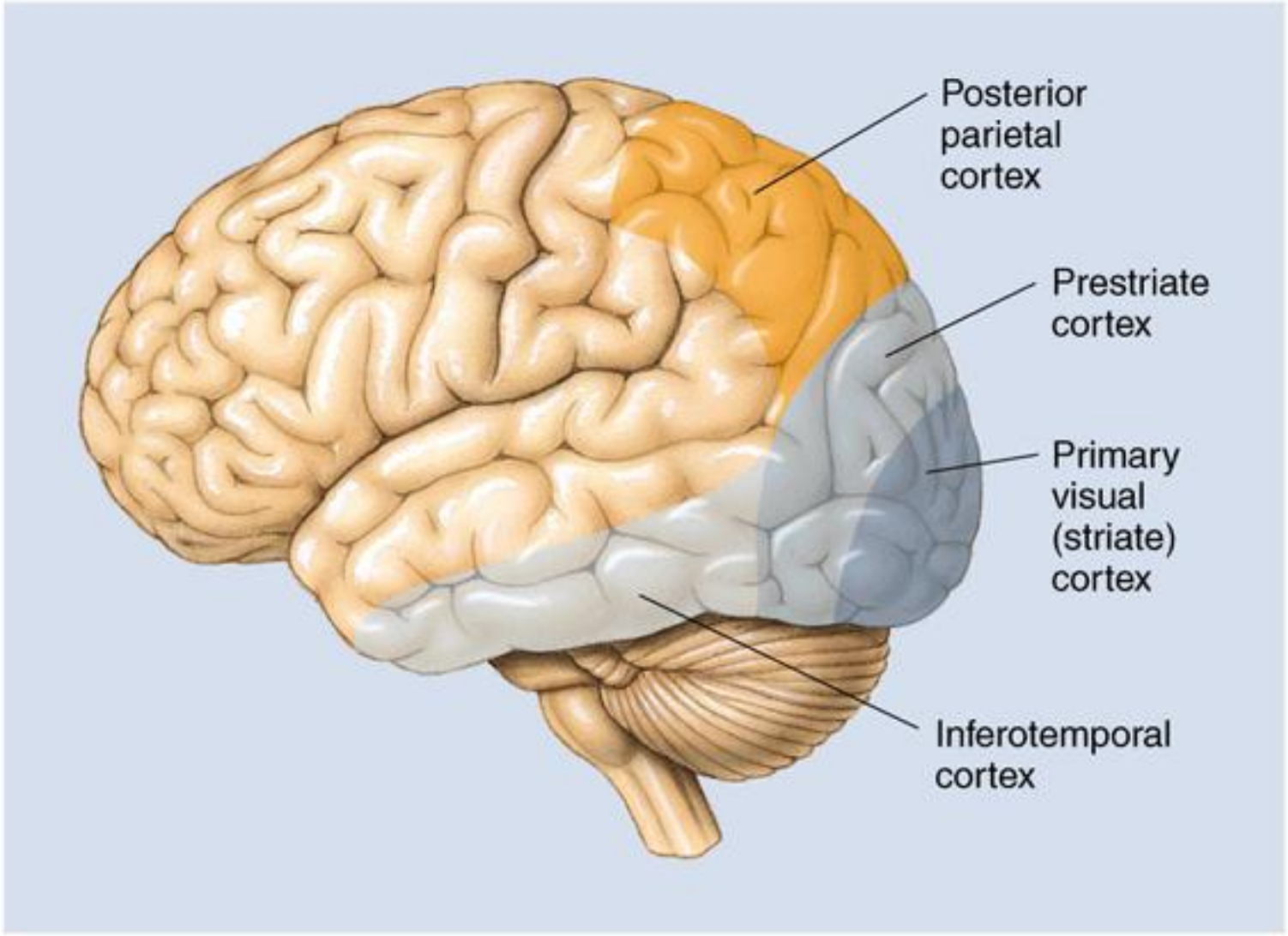
So What’s Going On?

Beyond V1 – Visual Association Cortex

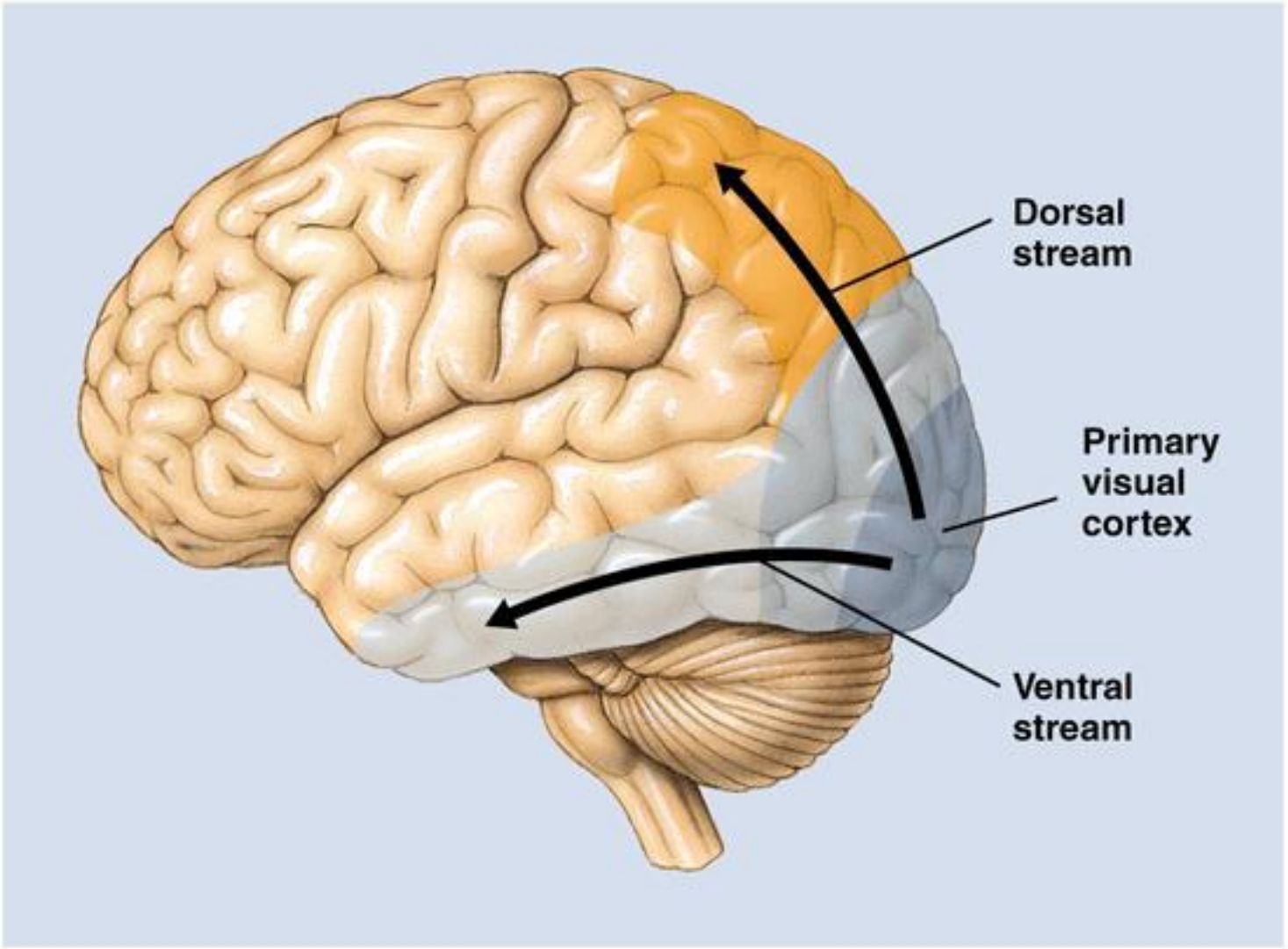
- “Serial” process to occipital cortex:

  Retina->Optic Nerve->LGN->Tracts->V1
- After V1 there are two major streams – Ventral “What” and Dorsal “Where” paths
- Functional segregation of vision – color, motion, faces.
- The “Binding” problem

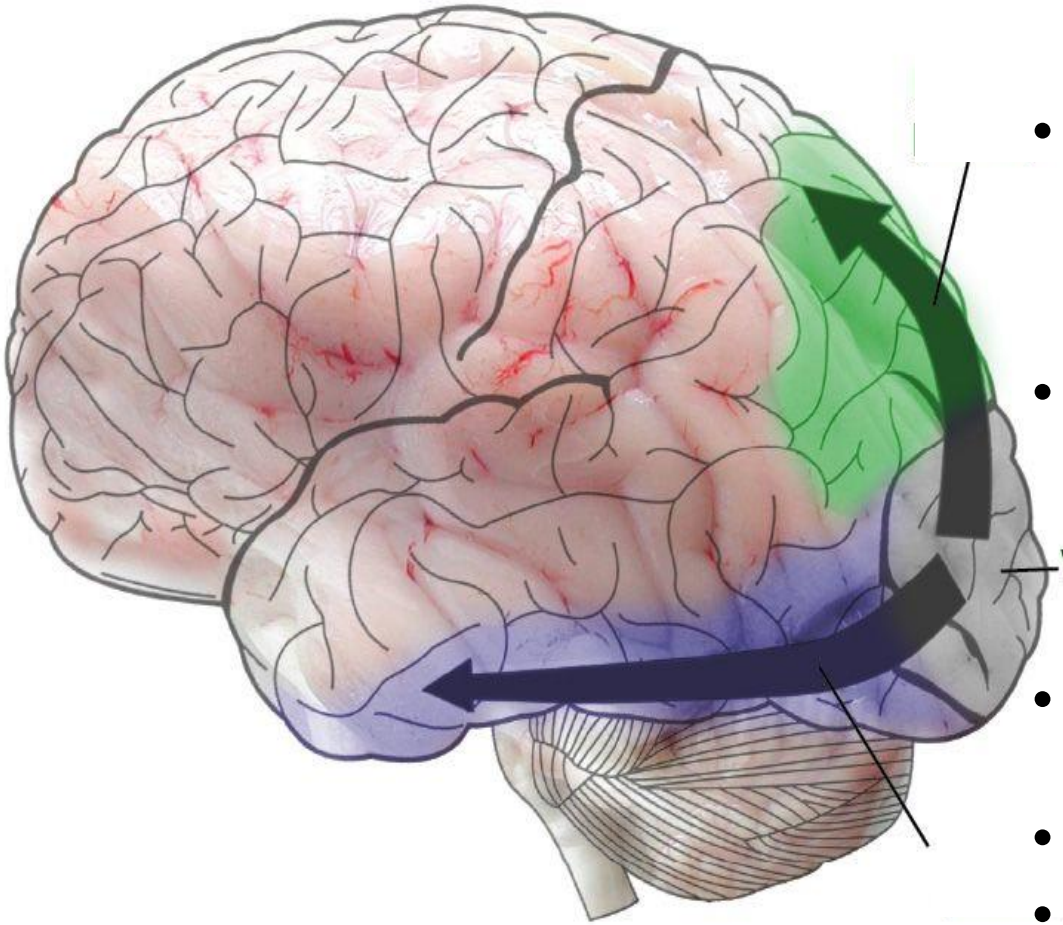
► Visual Areas of the Human Cerebral Cortex



► Visual Information Pathways

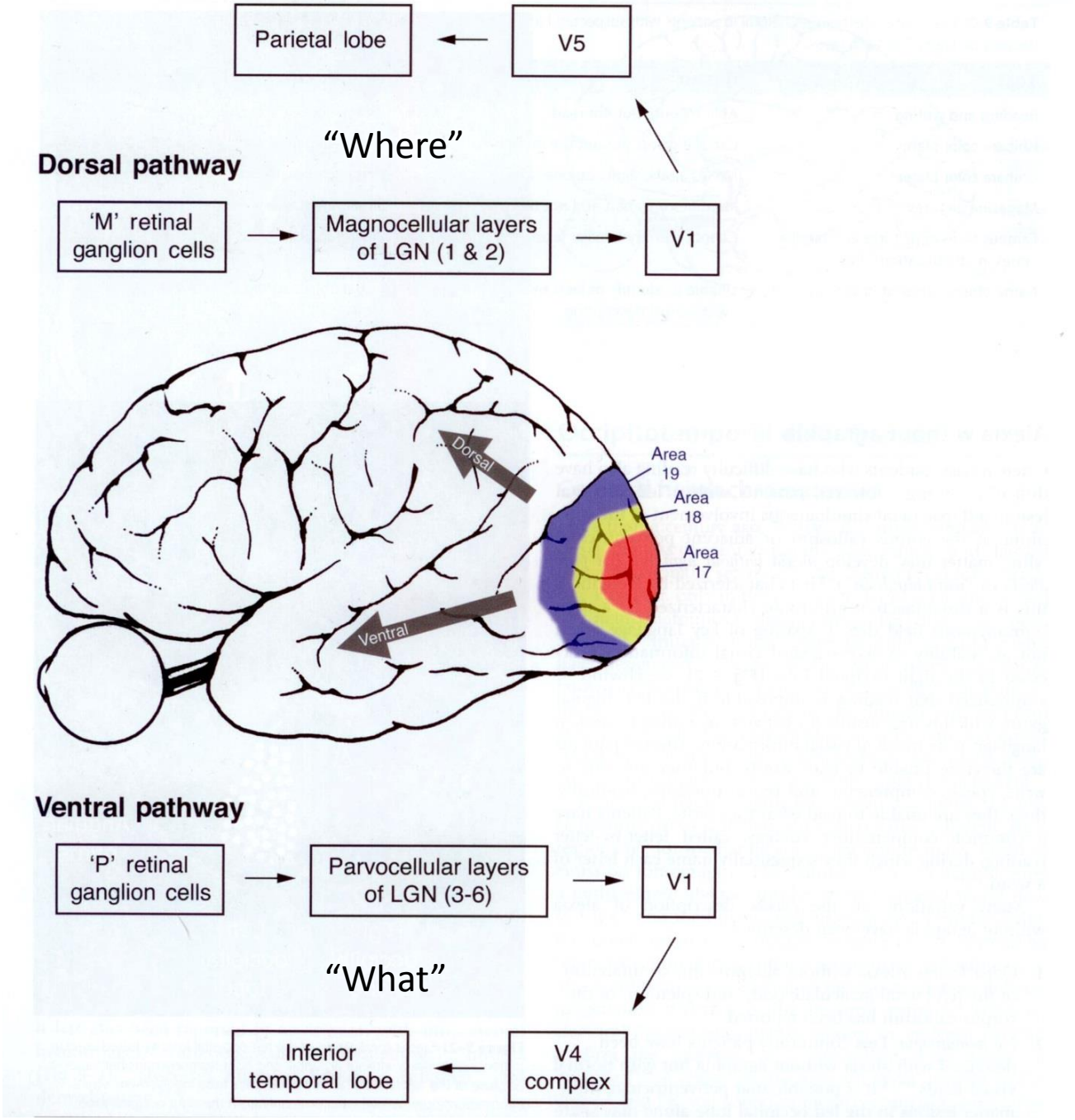


Two Stream Hypothesis



- Dorsal Pathway "Where" (magnocellular) → parietal lobe.
- Visual neglect

- Ventral Pathway "What"
- (parvocellular)
- → temporal lobe
- Visual agnosias.



From: *Neuro-Ophthalmology Diagnosis and Management* Liu, Volpe, Galetta

“What” Pathway

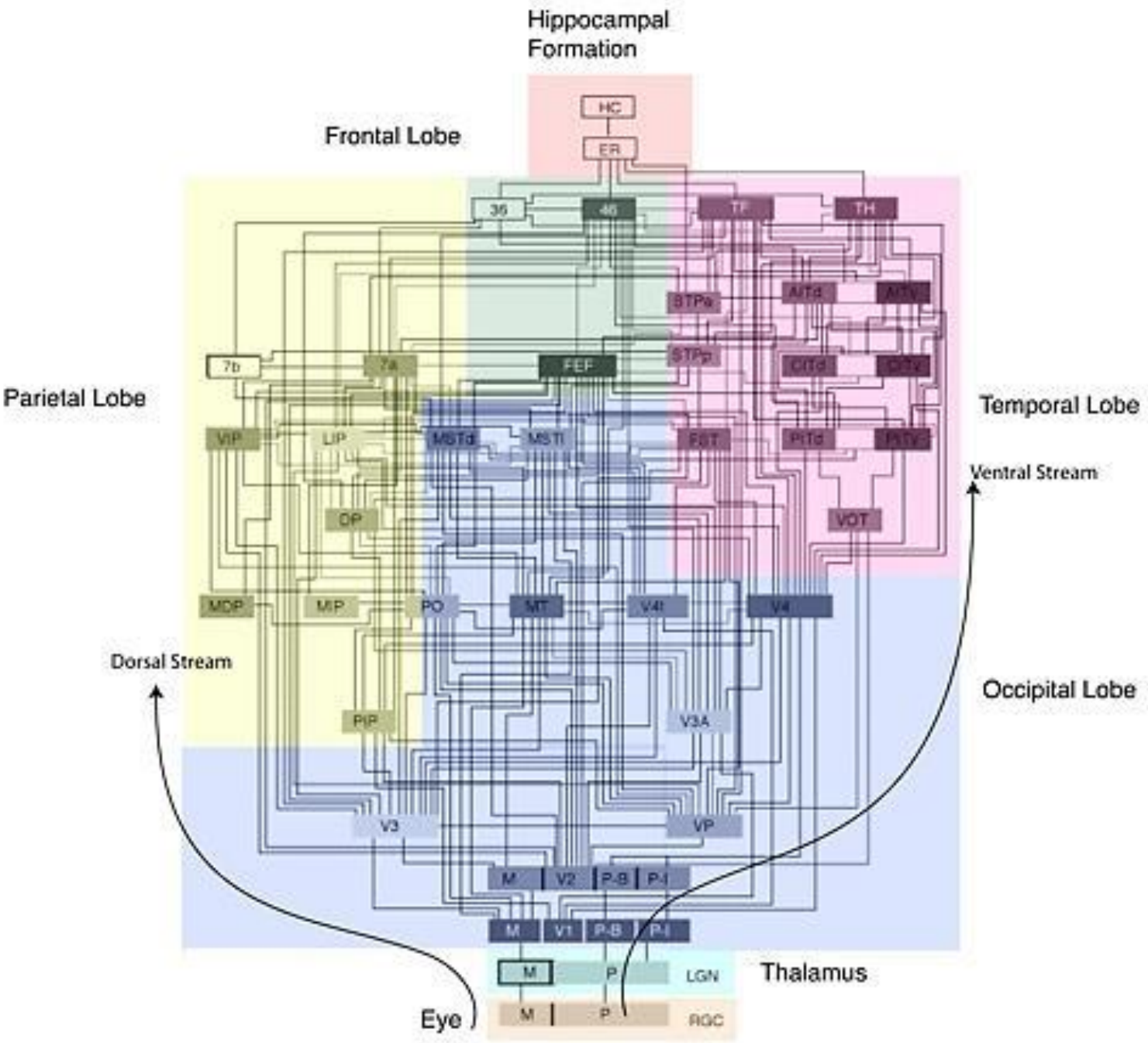
- Ventral stream (occipito-temporal) : object recognition, color, shape, form, and pattern.
- Continuation of the parvocellular pathway.
- V1→ V2→V4→ inferotemporal cortex→ angular gyrus→ limbic structures.
- Alexia, anomia, agnosia, amnesia.

“Where” Pathway

- Dorsal stream (occipitoparietal): Spatial orientation ,visual guidance of movement.
- Continuation of magnocellular pathway.
- V1→ V3→ V5→Parietal and superotemporal cortex.
- Simultanagnosia, anosagnosia, difficulty reaching for objects, acquired volitional movements of eyes, and hemispatial neglect (Balint’s syndrome).

Some Features of Visual Cortical Areas

- Mostly from monkey recordings, fMRI
- V1 – simple, complex & end stopped cells. Primary projections to V2 & V5
- V2 – orientation selective cells, half are color selective, convergence of input, larger receptive fields, depth perception selective
- V3/V3A – orientation selective cells, 50% color, 40% direction selective, binocularly driven
- V4 – shape, color, texture selective; preference for near, 50% color opponent; It's a color processing area
- V5 (MT) – 90% direction selective, MST has "motion in depth cells"; It's a motion processing area



http://vis.berkeley.edu/courses/cs294-10-fa07/wiki/index.php/A1-ArielRokem

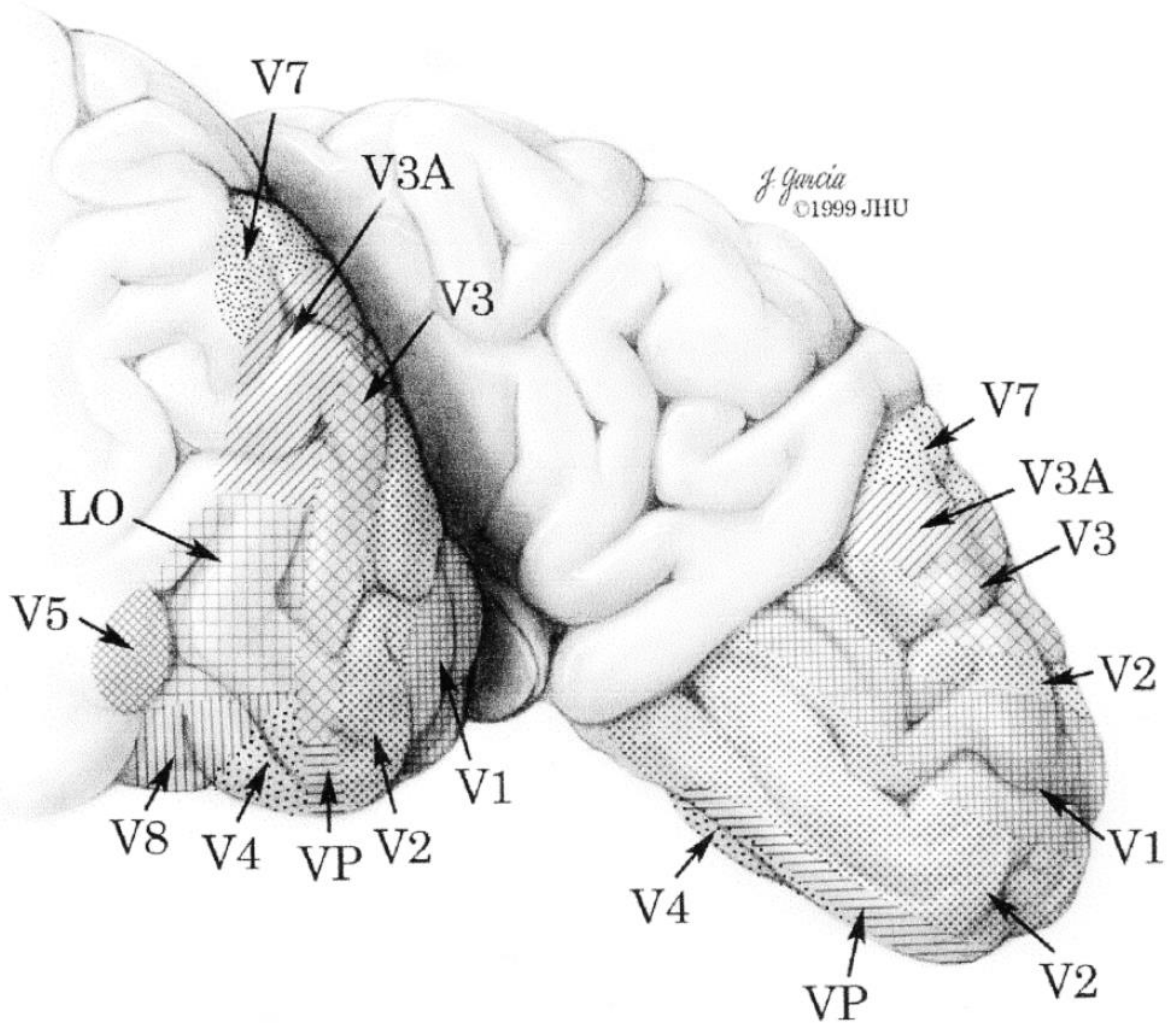


Fig. 3. Posterior lateral view of the human visual cortex showing several of the visual associative areas. The cerebellum has been removed and the hemispheres have been separated and displaced to display medial and lateral occipital regions. V1 corresponds to the primary of striate visual cortex. The other associative visual areas are discussed in the text except V7, V8, and LO (lateral occipital, which plays a role in object processing), because these areas have not been associated with distinct clinical syndromes. (Artist: Juan Garcia.)

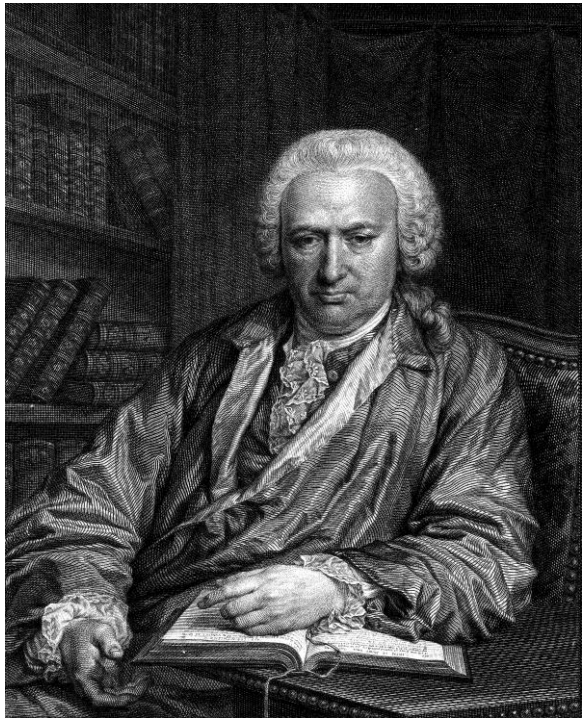
Selected Diseases of Visual Cortex

Cortical blindness

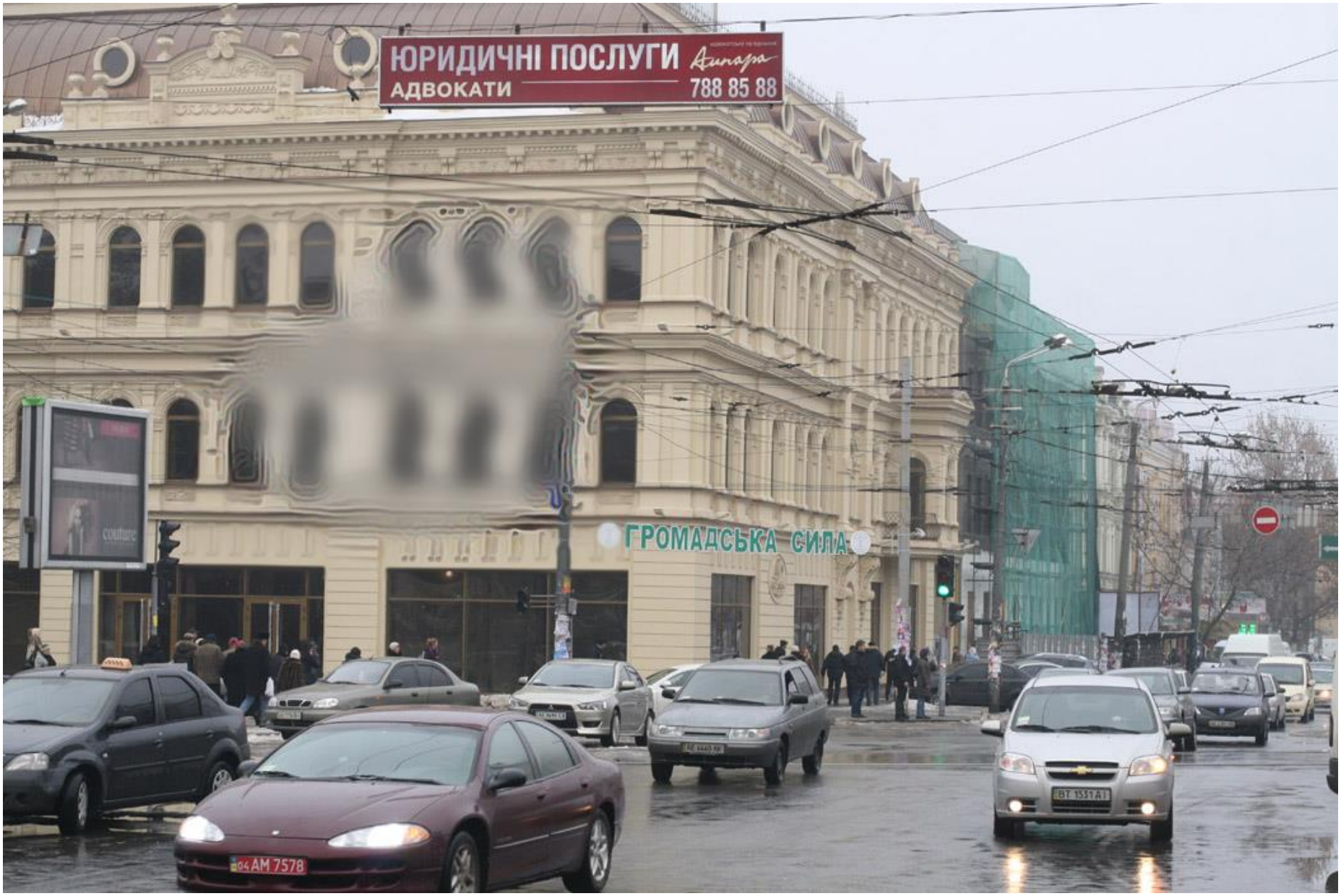
- Due to bilateral occipital lobe lesions.
- Sometimes misdiagnosed as functional vision loss.
- Causes: stroke, severe blood loss, eclampsia, hypertension, angiography, carbon monoxide poisoning.
- Anton’s syndrome

Release Hallucinations or Charles Bonnet Syndrome

- 1760 Swiss naturalist Charles Bonnet described hallucinations in his grandfather who had cataracts and vision loss.
- Hallucinations made worse by eye closure. There is disinhibition by the poor vision leading to the hallucinations. 80% of people with release hallucinations are over 60.
- Patients realize the hallucinations are not real but are relieved to hear they are “not going crazy”.
- Similar occurrence in hearing loss leading to musical hallucinations.
- Difficult to treat – usually reassurance is enough.



Migraine with Aura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Migraine Aura

- Wave of activation across visual cortex with a scotoma in its wake (spreading depression)

Alice in Wonderland Syndrome

Face Recognition

The Vegetable Gardener (1590)

Giuseppe Arcimbaldo (1526-1593)

Prosopagnosia

- Impaired ability to recognize familiar faces or learn new faces
- Use non-facial cues and are aware of their deficit
- Oliver Sacks: “The Man Who Mistook His Wife for a Hat”
- Location – Ventral stream -> bilateral inferotemporal cortex damage. Often associated with superior homonymous field defect.
  Can be a developmental defect.

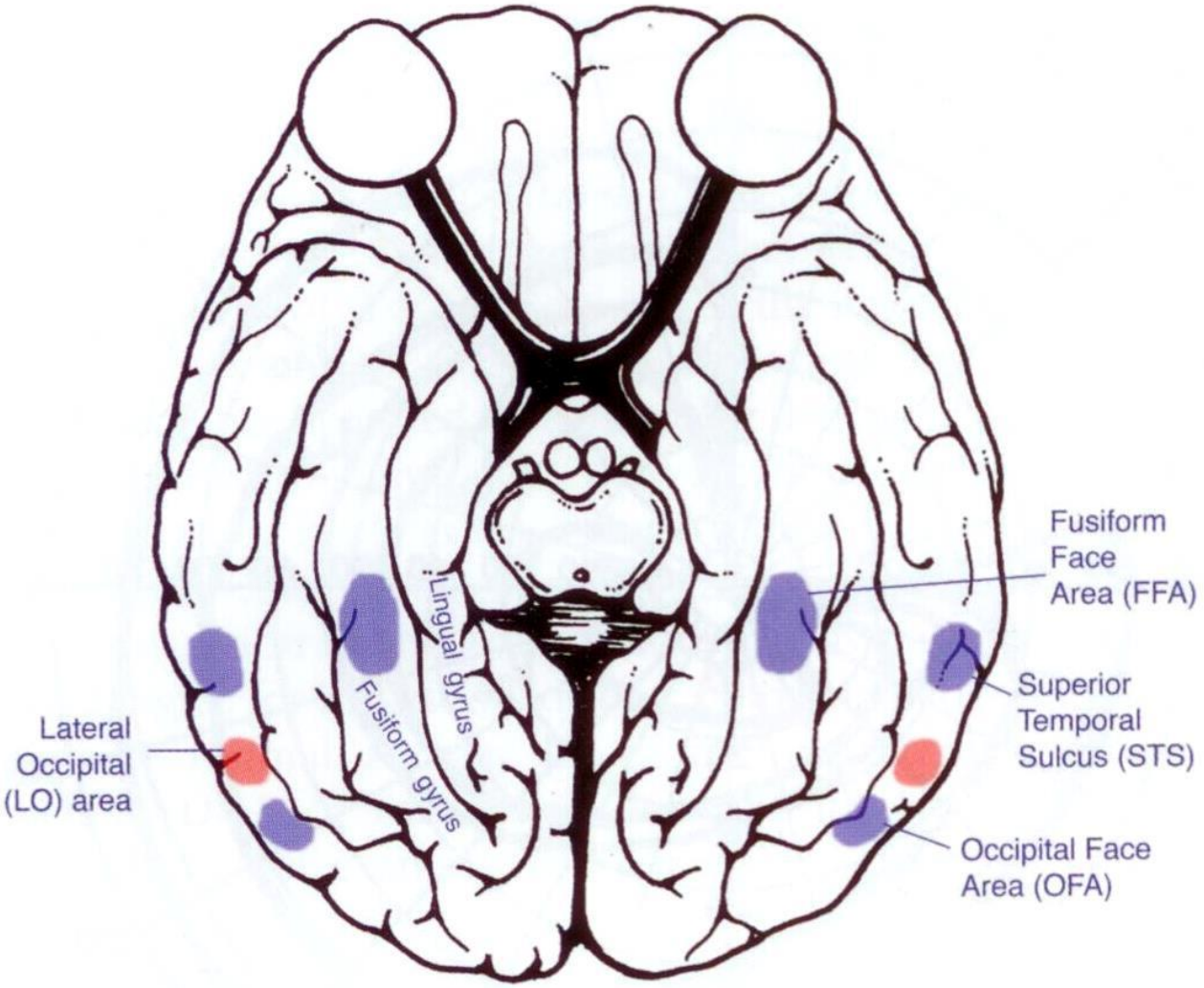


Figure 9–6. Underside view of the brain, highlighting the cortical areas active specifically during tasks of facial (FFA, STS, and OFA) and object recognition (LO).[105,116,168] Note V4 complex (not shown) lies more posteriorly in the fusiform gyrus (see Fig. 9–4). For simplicity, the highlighted areas, which are all bilateral, are labeled on only one side of the brain.

From: *Neuro-Ophthalmology Diagnosis and Management* Liu, Volpe, Galetta

Simultanagnosia

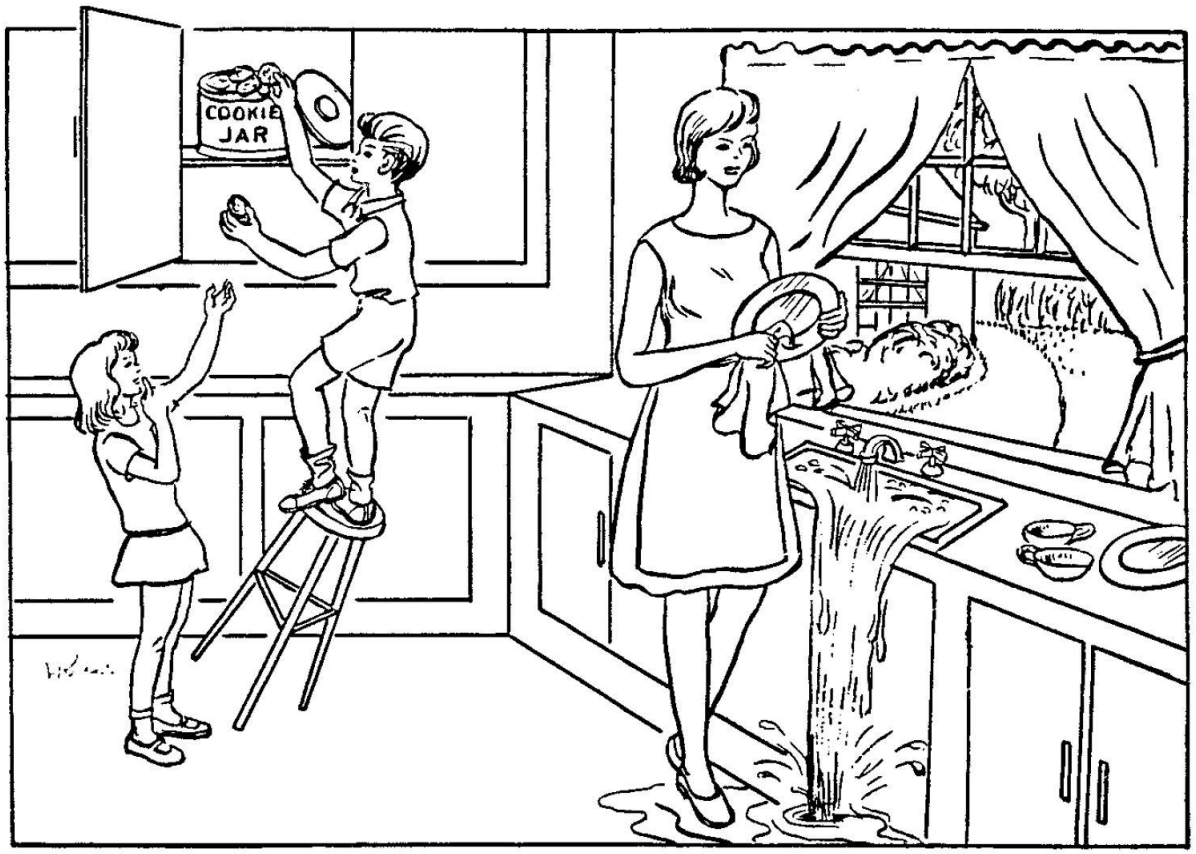
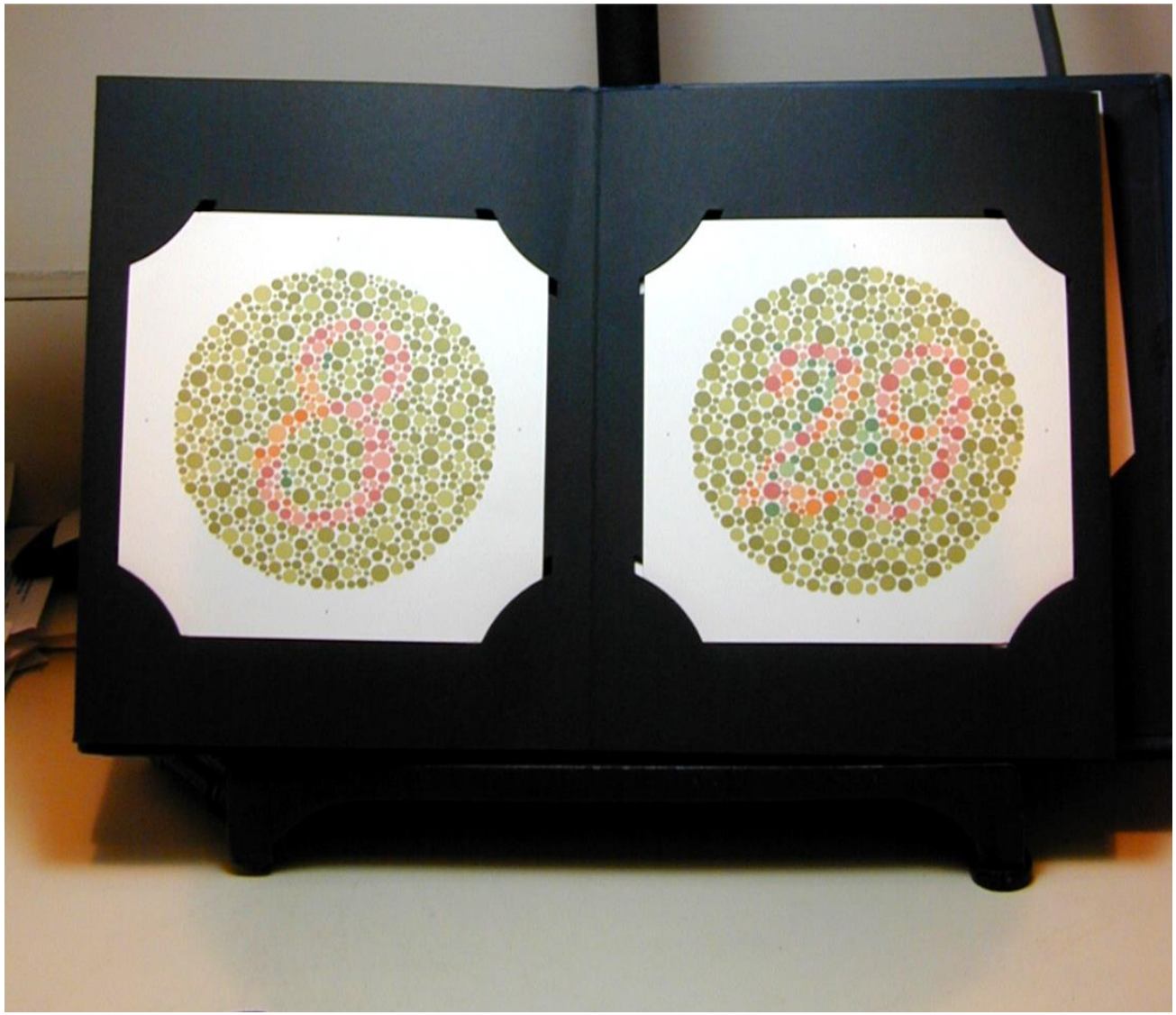


Figure 13.40. The Cookie Theft Picture from the Boston Diagnostic Aphasia Examination. This picture contains a balance of information among the four quadrants. The patient is asked to describe the events depicted in the picture.



Cerebral Dyschromatopsia

- Bilateral occipital lobe lesions in the lingual or fusiform gyri of the medial occipital lobe (medial occipito-temporal lobe – V4).
- Usually associated with a right superior quadrant or hemianopic field loss.
- Unilateral involvement may give hemidyschromatopsia.

Cerebral Dyschromatopsia

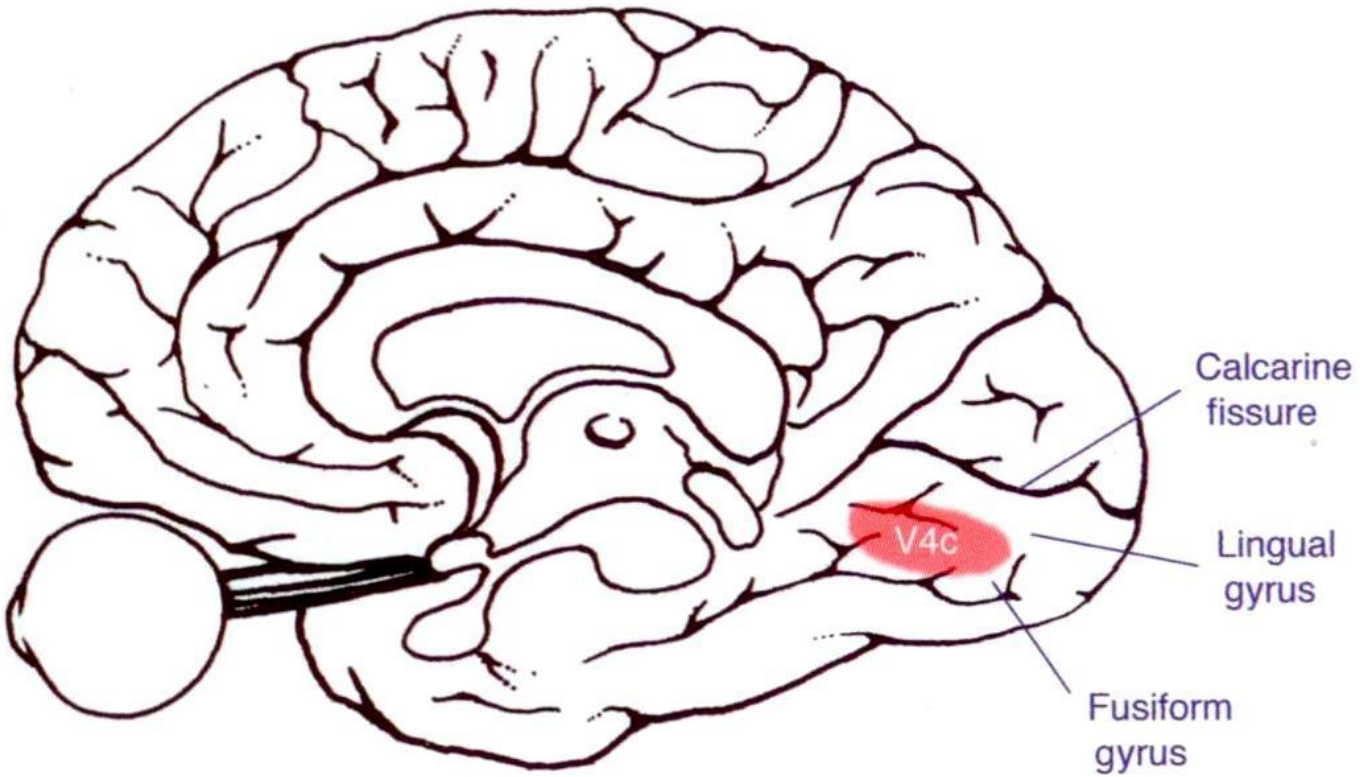


Figure 9–4. Medial view of the brain demonstrating location of area V4 complex, the human color center, which lies primarily in the posterior fusiform gyrus, with a lesser contribution from the lingual gyrus.

From: *Neuro-Ophthalmology Diagnosis and Management* Liu, Volpe, Galetta

Alexia without Agraphia

- Loss of ability to read but can write.
- Left occipital lobe and splenium of the corpus callosum involvement.
- Right homonymous hemianopia

Vision in the good left field (right occipital cortex) cannot get to the dominant angular gyrus (reading area) because the corpus callosum is also damaged.

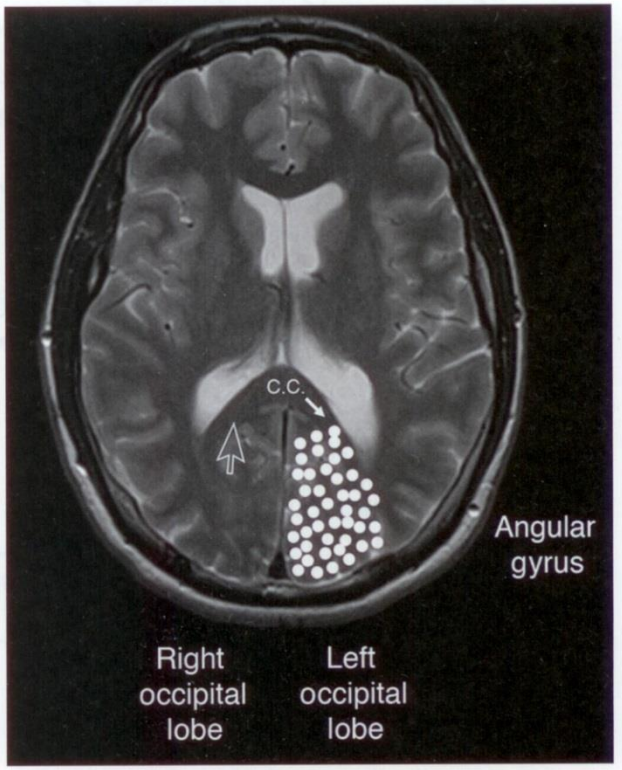


Figure 9–2. Classic localization of the left occipital lesion (dotted area) in patients with alexia without agraphia and right homonymous hemianopia. Because of the left lateral and mesial occipital lobe involvement, visual information (open arrow) can come only from the right occipital lobe. However, owing to disruption of the paraventricular white matter and the outflow of the corpus callosum (C.C.), this visual information (small arrow) cannot access the angular gyrus in the language areas.

From: *Neuro-Ophthalmology Diagnosis and Management* Liu, Volpe, Galetta

Palinopsia

- Perseveration of vision. Persistence or recurrence of visual images after the stimulus has been removed.
- Most commonly parieto-occipital damage with incomplete homonymous hemianopic visual field loss. The perseveration noted in the area of the field loss.
- Seen in migraine, certain drugs (LSD, trazadone, topirimate), epilepsy

Palinopsia

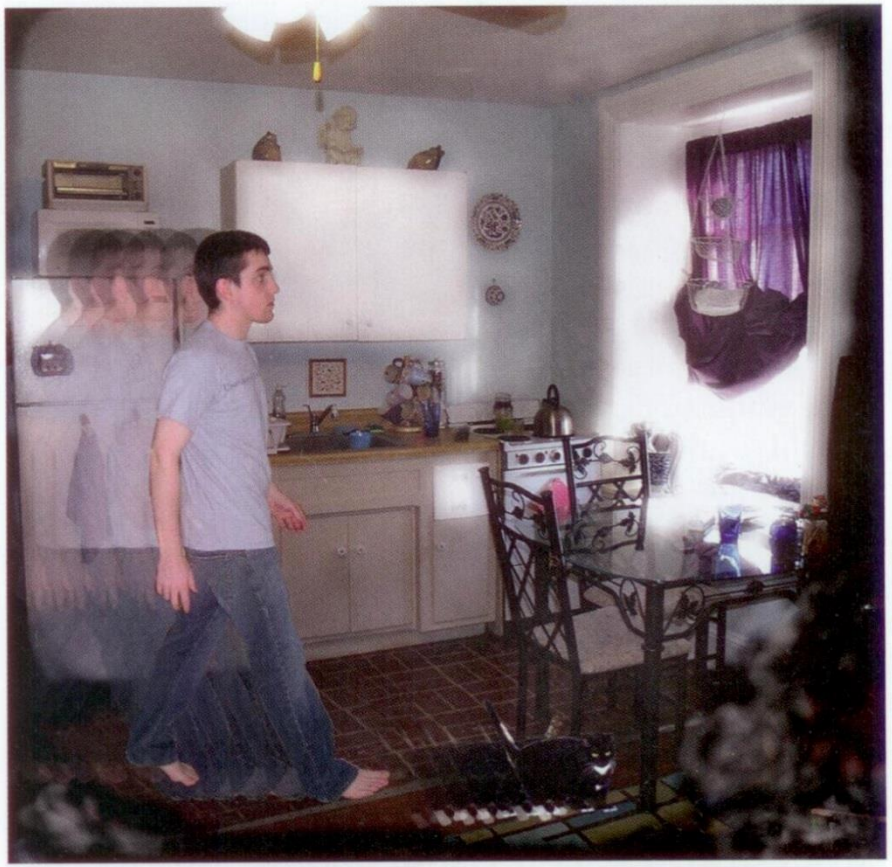


Figure 12–10. A patient's depiction of her palinopsia, which she describes as "a trailing effect behind moving objects" (the person and cat, for example).

From: *Neuro-Ophthalmology Diagnosis and Management* Liu, Volpe, Galetta

Other Unusual Visual Syndromes

- Visual neglect – patient ignores the left side of space (also left side of their body). -> Right inferior parietal lesion. (Abnormal clock drawing, line bisection)
- Micropsia, Macropsia – objects appear smaller or larger than normal. Can be seen in migraine.
- Akinetopsia – inability to see motion (bilateral occipito-temporal (V5) lesions). Patient LM

Treatment

- Sometimes can get recovery over time.
- Reassurance.
- Anticonvulsant and similar drugs can be tried but usually not helpful.
- PT, OT, adjustment over time

Sources

- *Walsh & Hoyt Clinical Neuro-Ophthalmology* Newman, Miller, Biousse, Kerrison. 6th Ed. Lippincott 2005 (available at NOVEL)
- *Neuro-Ophthalmology Diagnosis and Management* Liu, Volpe, Galetta. 2nd Ed. Saunders 2010

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J Neuro-Ophth


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Kennerdell, John S


Loewenfeld, Irene


Moran Eye Center


NANOS Annual Meeting


NANOS Historical Collection


Newman-Toker, D


Sadun, Alfredo A


Smith, J Lawton


Walsh & Hoyt Clinical Neuro-Ophth


Walsh Sessions


Wilhelm, Helmut


Wray, Shirley H

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